

Fatal toxic epidermal necrolysis-like lupus after longitudinally extensive transverse myelitis, vasculitic neuropathy and myositis despite reassuring serology: a case report

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Case Report

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Abstract

Background

Spinal cord and peripheral nerve involvement are among the most disabling manifestations of systemic lupus erythematosus, yet the disease activity index most used in routine practice has no descriptor for either, and anti-double-stranded DNA antibodies and complement are calibrated chiefly to nephritis. Toxic epidermal necrolysis-like lupus is a rare, life-threatening cutaneous manifestation that is usually confirmed by skin biopsy. To our knowledge, sequential spinal cord, peripheral nerve, muscle, marrow and skin involvement culminating in this complication has not previously been reported in a single patient.

Case presentation:

A 30-year-old Bengali woman with systemic lupus erythematosus and biopsy-confirmed lupus nephritis presented with seizure-like episodes, visual hallucinations and proximal lower-limb weakness. Within a month she developed longitudinally extensive transverse myelitis, severe axonal sensorimotor polyneuropathy, biopsy-proven vasculitic myositis and marrow hypoproliferation; aquaporin-4 and myelin oligodendrocyte glycoprotein antibodies were negative. Despite pulse methylprednisolone and six monthly cyclophosphamide pulses, she remained permanently paraplegic. Anti-double-stranded DNA antibodies fell below threshold once cyclophosphamide began, while anti-SSA/Ro, anti-Ro52 and anti-proliferating cell nuclear antigen antibodies remained strongly positive. Two months after induction ended, annular lesions progressed over 5 days to Nikolsky-positive epidermal detachment over approximately 55–60% of body surface area, with mild mucosal involvement. Apart from resumed maintenance mycophenolate mofetil, which she had previously tolerated, no drug had been introduced in the preceding 8 weeks; herpes simplex virus testing was negative, and dermatological assessment diagnosed toxic epidermal necrolysis-like lupus on clinical grounds because her condition precluded biopsy. Blood cultures grew multiple Gram-negative organisms and yeast, and she died within 48 hours despite only low-grade fever.

Conclusions

Normalising anti-double-stranded DNA antibodies and complement did not reflect this patient's progressive extrarenal disease, and her myelitis and neuropathy were not captured by the most widely used activity index. When skin biopsy is not feasible, toxic epidermal necrolysis-like lupus may be reached as a probable clinical diagnosis through formal drug-causality assessment, exclusion of infective mimics and attention to lupus-type morphology. Profoundly immunosuppressed patients with extensive skin loss may develop culture-proven sepsis with minimal systemic signs.

Background

Spinal cord and peripheral nerve involvement are among the most disabling manifestations of systemic lupus erythematosus (SLE), yet the instruments most used to follow the disease capture them poorly. The Systemic Lupus Erythematosus Disease Activity Index 2000 (SLEDAI-2K) has no descriptor for transverse myelitis or peripheral neuropathy [1], although both have American College of Rheumatology (ACR) case definitions [2]. The British Isles Lupus Assessment Group 2004 (BILAG-2004) index does include them [3] but is more demanding to complete, and the Systemic Lupus International Collaborating Clinics/ACR (SLICC/ACR) Damage Index records them only once a deficit has persisted for 6 months [4]. The serological markers most used to judge activity, anti-double-stranded DNA (anti-dsDNA) antibodies and complement, are calibrated chiefly to immune-complex nephritis.

Toxic epidermal necrolysis-like lupus erythematosus (TEN-like LE), part of the acute syndrome of apoptotic pan-epidermolysis (ASAP), is rare, affecting 6 of 9074 patients with cutaneous or systemic lupus in one large series [5, 6]. Distinguishing it from drug-induced toxic epidermal necrolysis (TEN) matters because management differs, and in most published cases the distinction was supported by skin biopsy [7]. We describe a patient in whom lupus moved sequentially through spinal cord, peripheral nerve, muscle, marrow and skin within 8 months while conventional serology normalised, ending in fatal TEN-like LE that had to be diagnosed without histology. To our knowledge, this sequence of organ involvement in a single patient has not previously been reported.

Case presentation

A 30-year-old Bengali woman with SLE and biopsy-confirmed lupus nephritis, diagnosed less than a year earlier and stable on hydroxychloroquine, prednisolone and mycophenolate mofetil, presented with seizure-like episodes, visual hallucinations, myalgias and proximal lower-limb weakness. Renal function remained normal throughout her illness. High-dose corticosteroids were started for neuropsychiatric SLE (NPSLE) with myelitis and myositis (see Investigations), and quetiapine was started for her psychiatric symptoms.

One month later she deteriorated, with asymmetric flaccid paraparesis (Medical Research Council grade 0/5 on the left and 2/5 on the right), absent knee jerks, diminished ankle jerks, complete urinary retention and pancytopenia.

Marrow function and right-leg power improved transiently by month 2, but paraplegia was permanent from month 3. Immobility and sphincter dysfunction led to recurrent culture-positive urinary tract infections with pyuria, including *Escherichia coli* and yeast at month 2, treated with co-trimoxazole, and to grade II–III pressure injuries.

Eight months after presentation, and 2 months after the final cyclophosphamide pulse, she was admitted with a 5-day eruption that began abruptly as annular erythematous lesions and progressed rapidly to Nikolsky-positive, sheet-like epidermal detachment (Fig. 1A). Detachment affected about 70% of the lower limbs and about half of the upper limbs, trunk and face, approximately 55–60% of body surface area; mucosal involvement was present but mild. Apart from mycophenolate mofetil maintenance

restarted after induction, no drug had been started in the preceding 8 weeks (see Differential diagnosis). She was cachectic, pale, icteric and tachypnoeic, with low-grade pyrexia (37.8°C), tachycardia and anasarca. Dense flaccid paraplegia (0/5) persisted with reduced touch sensation; cranial nerves were intact.

Investigations

At presentation, cerebrospinal fluid showed a lymphocytic pleocytosis with elevated protein, and culture was negative. Spinal magnetic resonance imaging (MRI) showed a longitudinally extensive intramedullary lesion from D4 to the conus medullaris with intramedullary enhancement (Fig. 1B–D), and thigh MRI showed active, left-predominant myositis (Fig. 1E). Brain MRI showed only mild diffuse cerebral atrophy. Serum anti-aquaporin-4 (AQP4) and anti-myelin oligodendrocyte glycoprotein (MOG) antibodies, tested before cyclophosphamide while she was receiving corticosteroids, were negative.

At the deterioration 1 month later, nerve conduction studies showed a severe axonal sensorimotor polyneuropathy, with myopathic changes on electromyography. Quadriceps biopsy showed lymphocytic vasculitis with perifascicular necrosis, myophagocytosis and vessel-wall thickening (Fig. 1F–I). Haemoglobin was 3.7 g/dL (transfused), platelets 45×10^9 /L, leukocytes 1.83×10^9 /L and reticulocytes 0.2%, consistent with severe marrow hypoproliferation; normal ferritin and triglycerides argued against macrophage activation syndrome. Antinuclear antibody was positive at 1:160, with anti-SSA/Ro, anti-Ro52, anti-RNP/Sm and anti-proliferating cell nuclear antigen (anti-PCNA) antibodies; anti-dsDNA was 18.09 IU/mL (threshold 10), C3 74.3 mg/dL, C4 52.3 mg/dL, erythrocyte sedimentation rate (ESR) 120 mm/hour and anticardiolipin IgG/IgM > 120 IU/mL. Anti- β 2-glycoprotein I antibodies were positive on repeat testing. SLEDAI-2K was 22 (seizure 8, vasculitis 8, myositis 4, low complement 2). Serial serology and haematology are shown in Tables 1 and 2.

Table 1
Serial serology by month after neuropsychiatric presentation

Marker	Month 1	Month 4	Month 6
Anti-dsDNA, IU/mL (threshold 10)	18.09	< 10	< 10
C3, mg/dL	74.3	116	39.1
C4, mg/dL	52.3	87.1	47.6
ESR, mm/hr	120	25	80
Anti-SSA/Ro, anti-Ro52, anti-PCNA	+++	+++	+++
Serum creatinine remained normal throughout (0.68 mg/dL at month 2; 0.79 mg/dL at month 4)			
Month 0 is the neuropsychiatric presentation; cyclophosphamide was given monthly from month 1 to month 6. anti-dsDNA, anti-double-stranded DNA; anti-PCNA, anti-proliferating cell nuclear antigen; ESR, erythrocyte sedimentation rate.			

Table 2
Serial haematological parameters by month after neuropsychiatric presentation

Parameter	Month 1	Month 2	Month 3	Month 4	Month 6	Month 8
Haemoglobin, g/dL	3.7 (transfused)	10	8.8	9.2	9.8	9.8
Leukocytes, $\times 10^9/L$	1.83	4.8	4.2	5.0	4.8	11.1
Platelets, $\times 10^9/L$	45	150	90	140	148	151
Month 0 is the neuropsychiatric presentation; month 8 values are from the terminal admission.						

At the terminal admission, leukocytes rose only modestly to $11.1 \times 10^9/L$ (neutrophil-predominant) from a baseline of $4.2\text{--}5.0 \times 10^9/L$ (Table 2), without high fever, alongside coagulopathy, hypoalbuminaemia and hyponatraemia. Blood cultures grew multiple Gram-negative organisms and yeast. Repeat autoimmune serology and skin biopsy were not performed because of her clinical instability.

Differential diagnosis

Myelitis. Infection was considered unlikely given negative cerebrospinal fluid culture. Because AQP4 antibody-positive neuromyelitis optica spectrum disorder (NMOSD) frequently coexists with SLE and anti-Ro positivity [8], NMOSD and MOG antibody-associated disease (MOGAD) were specifically tested for, and both antibodies were negative. Without AQP4 antibodies, a diagnosis of NMOSD requires at least two core clinical characteristics [8]; she had no optic neuritis or area postrema syndrome, brain MRI showed no demyelinating lesions, and myelitis was therefore her only core characteristic. MOGAD requires a positive MOG antibody [9]. The asymmetric deficit, coexisting axonal neuropathy and biopsy-proven vasculitis in muscle favoured vasculitic lupus myelopathy over demyelinating disease.

Epidermal detachment. Skin biopsy, which in most published cases supports the distinction between TEN-like LE and its mimics [7], was not possible, so the diagnosis rested on a structured exclusion of alternatives and on the clinical context.

Drug-induced Stevens-Johnson syndrome/TEN was the principal alternative. It does occur in patients with lupus [10], so it could not be assumed away. Every drug taken in the 8 weeks before onset was assessed with the Algorithm of Drug Causality for Epidermal Necrolysis (ALDEN) [11], under which a drug started more than 56 days before onset is an unlikely culprit. Hydroxychloroquine and prednisolone had been taken continuously for more than 8 months, quetiapine since the neuropsychiatric presentation 8 months earlier, a proton-pump inhibitor for years, and ibuprofen and aspirin intermittently for years without reaction. Cyclophosphamide had been started 7 months earlier and last given 2 months before onset. Co-trimoxazole, a drug with a well-recognised risk of epidermal necrolysis, had been given only for the urinary infection at month 2 and stopped about 6 months before onset. Mycophenolate mofetil, restarted after induction, had been taken for many months previously without any cutaneous reaction and is not recognised as a high-risk drug. No new drug had been introduced.

Infection-associated blistering was the second alternative. Staphylococcal scalded skin syndrome was a real concern in an immunosuppressed adult with recurrent bacterial infection, since like TEN it produces Nikolsky-positive detachment; however, it typically spares mucous membranes, which were involved here, and blood cultures grew Gram-negative organisms and yeast rather than *Staphylococcus aureus*. The bloodstream infection is more plausibly a consequence of extensive barrier loss, immobility and a urinary portal of entry than a trigger of the eruption. Erythema multiforme major associated with herpes simplex virus, and *Mycoplasma*-associated mucocutaneous eruption, were considered less likely: the eruption began as annular lupus-type lesions without target lesions, and herpes simplex virus testing was negative. *Mycoplasma* was not tested. Bullous SLE, which produces tense subepidermal blisters, did not fit sheet-like detachment.

Several features favoured TEN-like LE. The eruption began as annular lesions, a lupus-type morphology, before progressing to detachment, and dermatological assessment favoured TEN-like LE. She had established, highly active multiorgan SLE with persistently strong anti-SSA/Ro positivity, the autoantibody most often reported in TEN-like LE [7], and no temporally plausible drug trigger, which is among the most useful clinical clues to the diagnosis [7]. Her mild mucosal involvement does not discriminate, as mucosal involvement occurs in about half of reported TEN-like LE cases [7]. One feature did not fit: her detachment progressed over 5 days, whereas TEN-like LE typically evolves over days to weeks (median about 3 weeks) and drug-induced TEN over hours to days [7]. We therefore regard TEN-like LE as a probable diagnosis reached on clinical grounds, not a histologically proven one.

Treatment

High-dose corticosteroids were started at presentation. After the deterioration at month 1, pulse intravenous methylprednisolone was followed by oral prednisolone 50 mg/day for 1 week and 40 mg/day for 2 weeks before tapering, and by six monthly intravenous cyclophosphamide pulses of 1 g (months 1–6), with red-cell transfusion for anaemia. Mycophenolate mofetil was withheld during induction and restarted at its previous dose once the 6-month cyclophosphamide course was complete; hydroxychloroquine and low-dose prednisolone continued throughout. At the terminal admission she received broad-spectrum antimicrobials and supportive care; escalation of lupus-directed therapy was not possible because of the skin emergency and her rapid deterioration.

Outcome and follow-up

Outcome and follow-up

Paraplegia and sphincter dysfunction were permanent. Serology diverged from her clinical course (Table 1): anti-dsDNA fell below threshold once cyclophosphamide began and stayed undetectable, and C3 transiently normalised, while anti-SSA/Ro, anti-Ro52 and anti-PCNA remained strongly positive throughout.

At the terminal admission she deteriorated over 48 hours and died of polymicrobial Gram-negative and fungal septicaemia. Catastrophic antiphospholipid syndrome, for which infection is a recognised trigger, could not be excluded given her antiphospholipid antibodies and prolonged immobility. No autopsy was performed.

Patient perspective

From the patient's parents, translated from Bengali and read back to them.

Our daughter was doing well on her medicines for her kidney disease. Then, within a few months, everything changed. First she became confused, then she could not move her legs, and then she could not leave her bed at all. We did not understand how the same illness could suddenly attack so many parts of her body.

We live in a village far from Kolkata. Her treatment at the medical college was free, but travelling there again and again, and buying some of the medicines, was very hard for a family like ours. After she became paralysed, her husband left, and the two of us cared for her alone for most of a year.

Each time the doctors gave her stronger medicines, she would get a little better for a while and then worse again. This worried us greatly, because it seemed the medicines were no longer working on her. The doctors told us her illness was very severe and very rare. We could not understand all of it, but we understood that they were trying to keep her alive, and so were we. In the end her skin came off, and she died within two days. We still do not fully understand what took our daughter from us.

Discussion and conclusions

Her nephritis, the compartment most closely tracked by serology, stayed quiet throughout; recommendations from the European Alliance of Associations for Rheumatology (EULAR) caution that normal serology does not exclude active disease [12]. Instead, cord, nerve, marrow and finally skin became active in turn, while the instruments following her disease, composite activity scoring and anti-dsDNA/complement serology, stayed calibrated to a phenotype she had moved past.

Her seizures at presentation were captured by the neuropsychiatric descriptors of SLEDAI-2K, but the extension of the same flare a month later into cord, nerve and muscle was only partly captured: myositis and vasculitis have descriptors, whereas transverse myelitis and peripheral neuropathy do not [1], despite ACR case definitions for both [2]. The score therefore registered fragments without their anatomical extent. BILAG-2004 would have captured both [3], but SLEDAI-2K is the simpler and more widely used index in routine care. The SLICC/ACR Damage Index records both, but only once a deficit has persisted for 6 months [4], which is what her paraplegia became. Each index worked as designed, yet between SLEDAI-2K and the Damage Index lay an interval in which neurological disease could be active without registering in either. The SLEDAI-2K vasculitis item, with its site-unrestricted biopsy

clause, could register her quadriceps findings, but it scores a fixed 8 points regardless of site and cannot distinguish a self-limited infarct from vasculitis destroying the cord.

Her serology then turned from confirmation to concealment. Anti-dsDNA and C3 track immune-complex nephritis, and cyclophosphamide suppressed this axis; anti-SSA/Ro, anti-Ro52 and anti-PCNA have no equivalent monitoring role, yet stayed strongly positive throughout. ESR, which lies outside this axis, fell alongside anti-dsDNA at month 4 and rose again by month 6, a non-specific signal against otherwise reassuring serology. C3 fell substantially by month 6 while C4 stayed relatively preserved, so complement did not parallel her deterioration, illustrating its limited sensitivity for non-renal disease.

The epidermal event occurred during the transition from induction to maintenance: the marrow effects of cyclophosphamide outlast the final pulse, and mycophenolate mofetil, restarted after induction, may not yet have been fully protective. One speculative explanation for the eruption is re-emergence of an autoreactive, Ro52-associated apoptotic pathway [13]; persistent anti-SSA/Ro and anti-Ro52 positivity is plausible context, not proof of causation. Neither anti-dsDNA positivity nor low C4, the features most associated with systemic involvement in TEN-like LE [7], was present when her epidermis failed. Her antiphospholipid antibodies, high-titre anticardiolipin and anti- β 2-glycoprotein I, may have contributed, although without a documented thrombotic event classification criteria for antiphospholipid syndrome were not met.

In a 2025 systematic review of 112 published cases of TEN-like LE, about half arose in patients with pre-existing lupus, and 6 patients (5.4%) died, 3 of them from sepsis [7]. Most of those cases included a skin biopsy. When histology is unavailable, as here, the diagnosis depends on a structured clinical approach: formal drug-causality assessment, deliberate exclusion of infective mimics, and attention to lupus-specific clinical clues such as annular onset.

Three factors made her terminal sepsis hard to recognise by ordinary criteria: impaired marrow reserve; months of corticosteroid and alkylating therapy blunting the host response; and loss of more than half of her skin barrier, removing both physical defence and physiological alarm. Despite culture-proven polymicrobial bacteraemia and fungaemia, she had only low-grade fever and a modest leukocyte rise. The case has important limitations. Attribution of her neurological disease to vasculitic lupus is inferential, since neural tissue was never biopsied; the skin diagnosis is clinical, without histology or immunofluorescence; catastrophic antiphospholipid syndrome could not be excluded without autopsy; and the mechanisms proposed above are hypothesis-generating.

Conclusions

This case carries five practical lessons. First, once anti-dsDNA and complement have normalised on induction therapy, persistently positive anti-SSA/Ro, anti-Ro52 or anti-PCNA antibodies, or a rising ESR, should not be read as reassurance of quiescence. Second, SLEDAI-2K has no descriptor for active transverse myelitis or peripheral neuropathy, and the SLICC/ACR Damage Index records them only as fixed damage after 6 months; BILAG-2004 or serial clinical examination is needed to capture them. Third,

the transition from cyclophosphamide induction to maintenance immunosuppression is a period of vulnerability that warrants close monitoring. Fourth, in marrow-suppressed, barrier-compromised patients with lupus, even culture-proven polymicrobial sepsis may produce only low-grade fever and a modest leukocyte rise, so suspicion should rest on anatomical and pharmacological risk. Finally, rapidly progressive epidermal detachment in SLE warrants formal drug-causality assessment and consideration of TEN-like LE, which may have to be diagnosed on clinical grounds when skin biopsy is not feasible.

Abbreviations

ACR

American College of Rheumatology

ALDEN

Algorithm of Drug Causality for Epidermal Necrolysis

anti-dsDNA

anti-double-stranded DNA

anti-PCNA

anti-proliferating cell nuclear antigen

anti-RNP/Sm

anti-ribonucleoprotein/Smith

anti-SSA/Ro

anti-Sjögren's syndrome-related antigen A/Ro

AQP4

aquaporin-4

ASAP

acute syndrome of apoptotic pan-epidermolysis

BILAG-2004

British Isles Lupus Assessment Group 2004 index

ESR

erythrocyte sedimentation rate

EULAR

European Alliance of Associations for Rheumatology

H&E

haematoxylin and eosin

IgG/IgM

immunoglobulin G/immunoglobulin M

LE

lupus erythematosus

MOG

myelin oligodendrocyte glycoprotein

MOGAD

MOG antibody-associated disease

MRI

magnetic resonance imaging

NMOSD

neuromyelitis optica spectrum disorder

NPSLE

neuropsychiatric systemic lupus erythematosus

SLE

systemic lupus erythematosus

SLEDAI-2K

Systemic Lupus Erythematosus Disease Activity Index 2000

SLICC

Systemic Lupus International Collaborating Clinics

STIR

short tau inversion recovery

TEN

toxic epidermal necrolysis

Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

Written informed consent was obtained from the patient's next of kin (her father) for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests

The authors declare that they have no competing interests.

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Author Contribution

AL assisted in managing the patient, collected the clinical data and drafted the manuscript. NR handled the administrative aspects of the case and assisted in manuscript preparation. Both authors reviewed and approved the final manuscript. AL is the guarantor.

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Availability of data and materials

Data sharing is not applicable to this article as no datasets were generated or analysed during the current study.

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Figures

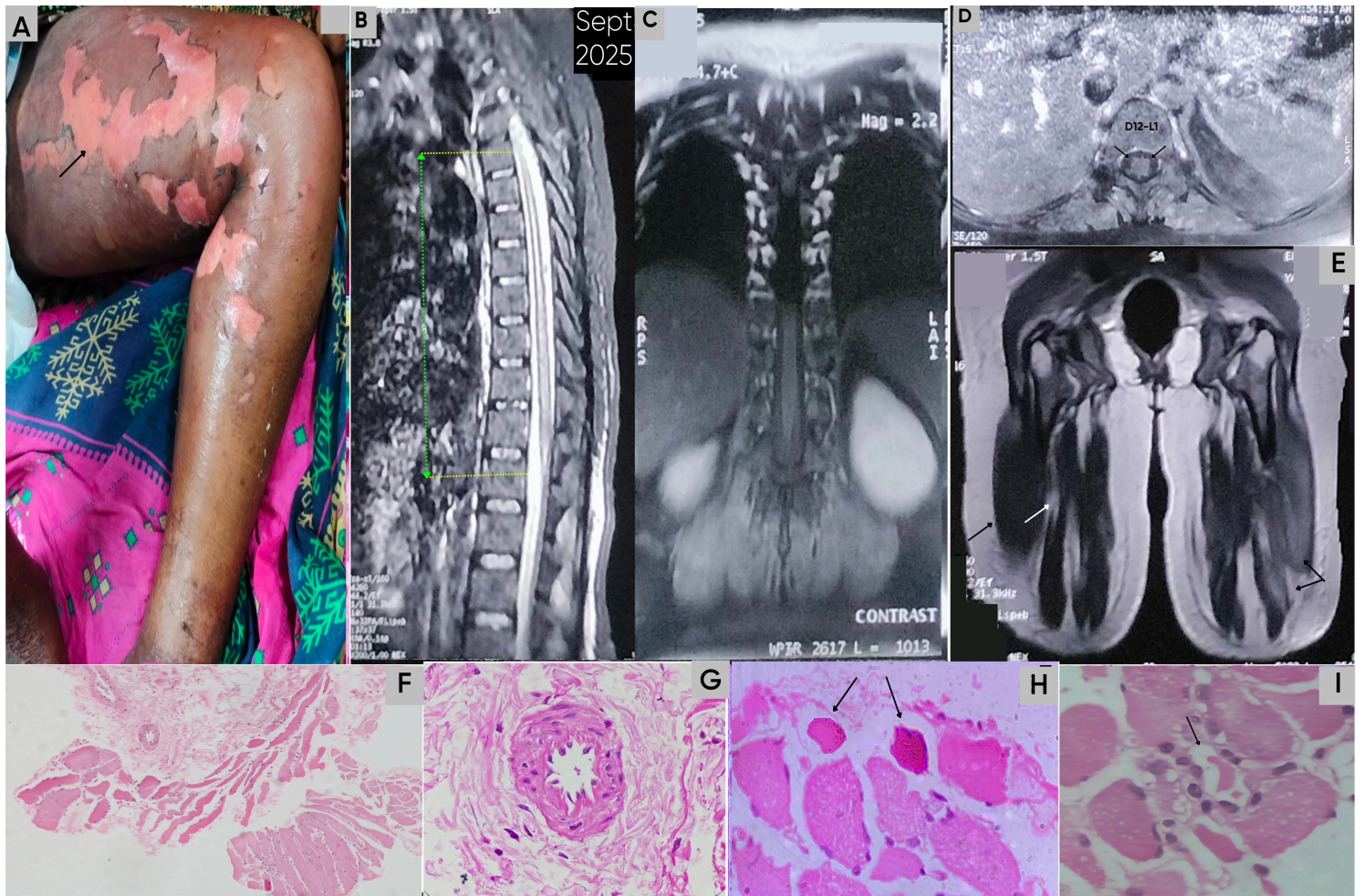


Figure 1

Multisystem manifestations culminating in fatal toxic epidermal necrolysis-like lupus

(A) Sheet-like epidermal detachment with Nikolsky-positive advancing margin (arrow). (B) Sagittal STIR MRI: diffuse intramedullary hyperintensity from D4 to the conus (dotted lines), consistent with longitudinally extensive transverse myelitis. (C–D) Coronal and axial post-contrast T1-weighted images at D12–L1: intramedullary enhancement (arrow), consistent with disruption of the blood–spinal cord barrier. (E) Coronal STIR MRI of the thighs: asymmetric, left-predominant myositis (black arrow), septal oedema (white arrow) and anasarca. (F) Disrupted fascicular architecture, myonecrosis and interstitial mononuclear infiltration. (G) Lymphocytic vasculitis: circumferential mononuclear infiltration, mural thickening and luminal narrowing. (H–I) Perifascicular necrosis (arrows) and myophagocytosis (arrow), supporting microvasculopathic ischaemia. H&E; F ×45, G–I ×100.

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